

# Spontaneous intramural oesophageal perforation

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Two cases of spontaneous intramural perforation of the oesophagus are described. In both, the area of dissection was extensive. The former recovered without operation; the latter required right thoracotomy and mediastinal and bilateral pleural drainage. Functionally, both have subjectively normal swallowing, but with fluoroscopic and manometric depression of peristalsis.

In the light of this recent clinical experience the classification of oesophageal perforation is redefined.

The differences between spontaneous *intramural* and *transmural* perforation of the oesophagus are stressed, as well as the importance of recognizing this new form of oesophageal perforation as a further lesion in the differential diagnosis of sudden, severe, retrosternal pain.

To date, both 'acquired' and 'spontaneous' perforations of the oesophagus have been described. Acquired perforations have further been encountered as 'transmural' or 'intramural' defects.

The purpose of this paper is both to describe two cases of *spontaneous* intramural perforation of the oesophagus—the second and third on record—and to redefine the classification of oesophageal perforations (Table).

TABLE

A. Acquired Perforation
(1) Acquired Transmural Oesophageal Perforation (ATOP)
(2) Acquired Intramural Oesophageal Perforation (AIOP)
B. Spontaneous Perforation
(1) Spontaneous Transmural Oesophageal Perforation (STOP)
(2) Spontaneous Intramural Oesophageal Perforation (SIOP)

## ACQUIRED TRANSMURAL OESOPHAGEAL PERFORATION

For long this has been well recognized as a complication of oesophageal foreign bodies, of their removal by oesophagoscopy, or rarely from oesophagoscopy inspection or treatment of benign or malignant oesophageal strictures (Overstreet and Ochsner, 1955; Borrie, 1958).

## ACQUIRED INTRAMURAL OESOPHAGEAL PERFORATION

Acquired intramural perforation resulting in an abscess as a complication of foreign bodies was described as a localized lesion simulating carcinoma by Sanborn (1960). He also referred to the acute localized 'oesophageal phlegmons', de-

scribed by the earlier endoscopists, which undoubtedly were localized acquired intramural perforations by foreign bodies with intramural abscess formation.

Such acquired intramural perforations can also be caused by an instrument and may be accompanied by extensive dissection within the loose submucosal layer, thus separating the mucosa from the muscular wall (Borrie, 1958; Lichter and Borrie, 1965). They usually occur in the upper third of the oesophagus, especially in the cervical oesophagus. The mucosal breach allows saliva and food to gain access to the loose submucosa, thereby extending the size of the false passage and producing a cylindrical dissection with extensive separation of the mucosa from the surrounding oesophageal muscular wall. This stripping may extend from the origin of the oesophagus at least as far as the aortic arch. The radiological finding of a 'double-barrelled oesophagus' with radio-opaque dye, such as sodium diatrizoate (Gastrografin), is diagnostic of this lesion.

No age seems immune, for Eklöf, Löhr, and Okmian (1969) have recently described three cases in neonates caused by the trauma of oral and pharyngeal suction with a rigid catheter.

## SPONTANEOUS TRANSMURAL OESOPHAGEAL PERFORATION

Though historically the oldest type recognized, and vividly described in 1724 by Boerhaave (Barrett, 1946), it was not until 1947 that Barrett

reported the first successful surgical repair. Early recognition of the perforation—which may extend into either or both pleural cavities, or the mediastinum alone—has enabled prompt thoracotomy with suture to become standard practice for this thoracic surgical emergency (Borrie, 1958).

However, a recent review article on spontaneous oesophageal perforation (Leading article, 1969) did not mention spontaneous *intramural* oesophageal perforation, a case of which had earlier been described from Capetown by Marks and Keet (1968).

#### SPONTANEOUS INTRAMURAL OESOPHAGEAL PERFORATION

In 1967 we first encountered this variant as a new phenomenon. In contrast to the former, it occurred in the lower third of the oesophagus. Like the first case reported by Marks and Keet (1968), it was recognized from the history and characteristic radiographs, and was successfully treated without operation.

#### CASE REPORTS

**CASE 1** Mr. R., No. 104589, aged 47 years, was admitted to Wakari Hospital as a medical emergency on 22 September 1967. Twenty years earlier he had had a terminal ileostomy for ulcerative colitis followed in 1960 by a total colectomy.

On 21 September, while in the very act of swallowing a portion of sandwich lunch, he stifled a sneeze and was immediately aware of retrosternal discomfort. He had never experienced this before. Some discomfort persisted throughout the afternoon and at 5.30 p.m. he drank a glass of milk. This was immediately followed by severe epigastric pain that came in waves lasting 5–6 seconds, occurring every 30 seconds for over an hour. He then retched up, once only, some 20 ml. of fresh blood. He presently noticed a black discharge from his ileostomy.

Throughout the ensuing night he was nauseated, and a 'raw feeling' persisted in the epigastrium. The next morning he was admitted to hospital.

**Examination** This revealed no abnormality other than mild distress from his deep-seated epigastric

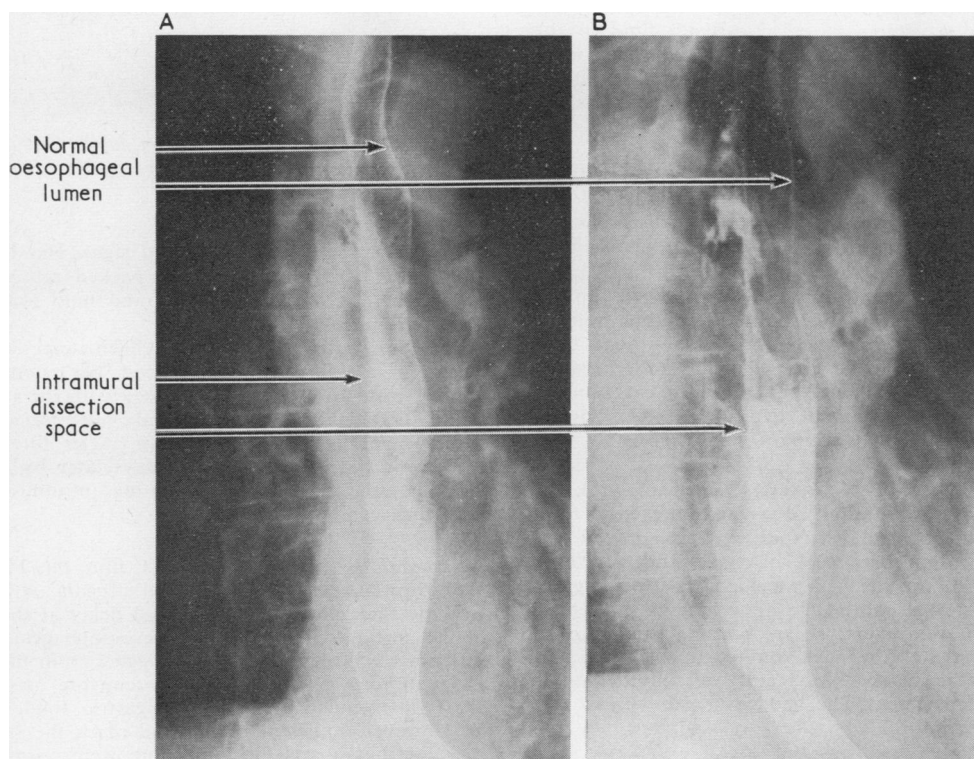


FIG. 1. Case 1. Barium swallow on day 4 shows a 'double-barrelled oesophagus' extending from the level of the aortic arch to the cardia.

ache. His temperature was 37.5° C., pulse 88 per minute, respirations 20 per minute; his packed cell volume was 34% and the electrocardiogram was normal. He was placed on a 'gastric regime' and observed. Within 30 hours his temperature had returned to normal. He continued taking a fluid diet.

**Investigations** A barium swallow performed four days after the onset showed a 'double-barrelled oesophagus' extending from the level of the aortic arch to the cardia (Fig. 1). The site of entry was not conclusively demonstrated, but, as a delayed film showed no remnants of barium in the mediastinum, it was presumed that dependent drainage was occurring. Nowhere was the oesophageal musculature breeched, nor was there any sign of mediastinal or cervical emphysema. The stomach and duodenum appeared normal.

This lesion was recognized as an 'intramural perforation of the oesophagus' with dissection in the submucous plane.

**Management** He was referred for a thoracic surgical opinion nearly a week after onset. At this time he was reasonably well and taking a soft diet. However, in view of the radiological extent of the mucosal dissection, it was felt reasonable to stop oral feeding and begin a regime of caval catheter fluid for one week. Since adequate drainage of the space in the oesophageal wall had been demonstrated radiologically, any surgical procedures seemed pointless, and oesophagoscopy was not indicated.

**Progress** Subsequent radiological examinations were made on 29 September and 5 October. A small bulge was shown on the left posterolateral aspect of the oesophagus a little above the cardia at one of the usual sites of total perforation (Fig. 2). The mucous membrane appeared reattached to the muscle layer and the oesophageal outline returned to normal. However, peristalsis was absent below the level of the aortic arch. On 5 October 1967, as his symptoms had gone, he started to eat a normal diet and was discharged two days later. He has remained well and working.

**CASE 2** Mrs. M., No. 800724, aged 65, a known hypertensive, was admitted as an emergency on 10 May 1969. She had been perfectly well in the early evening, eating her meal of steak and vegetables. About 9.30 p.m. she had a sudden onset of epigastric and retrosternal pain at the same time as she retched. This pain was severe, sharp, burning in nature and did not radiate. She next vomited up dark fluid followed by bright red blood and was admitted to the medical department. She had never experienced these symptoms before.

**Examination** This revealed a thin woman frequently vomiting blood. Her temperature was 37° C., pulse 98 and blood pressure 150/85 mm. Hg.

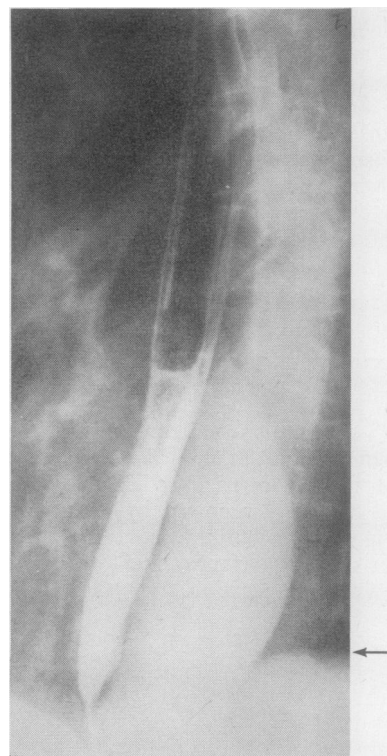


FIG. 2. Case 1. Barium swallow on day 8. Left anterior oblique projection shows a small bulge on the left posterolateral aspect of the oesophagus.

There were no abnormal physical signs. Her haemoglobin was 10.8 g./100 ml. and packed cell volume 34%. The electrocardiogram showed mild ischaemic change.

She was initially treated as 'haematemesis' and, when given 1 litre of whole blood, her packed cell volume rose to 40%. She continued with intravenous fluids. Next day she still had steady retrosternal pain, made worse by swallowing saliva. After three days she drank water, followed two days later by a fluid diet. She received ampicillin, 500 mg. intramuscularly four times daily.

**Investigations** A frontal chest film on 11 May 1969 showed a small right pleural effusion. A barium swallow the following day showed delay at the level of the aortic arch. Below this the oesophageal lumen was not easily defined, being deflected from the right and anteriorly by a long space-occupying mass (Fig. 3). This extended down to the gastric fundus (Fig. 4). However, sufficient barium did reach the stomach to show that it and the duodenum were normal. No barium entered the mediastinum. The appearances suggested partial oesophageal rupture with intramural dissection.

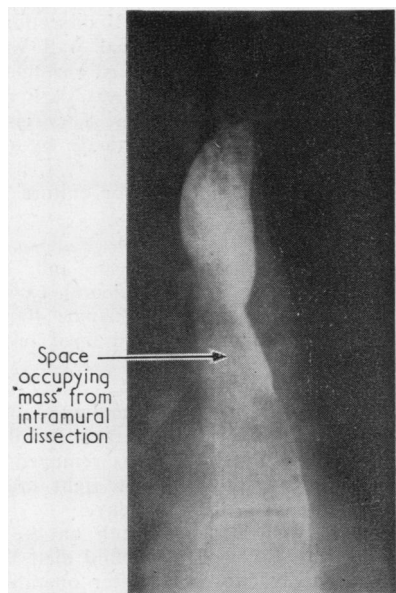


FIG. 3. Case 2. Barium swallow on day 2 shows extensive deflection of oesophageal lumen from the right side by a long space-occupying mass.

*Initial progress* She developed a nightly swinging temperature of  $38.5^{\circ}\text{C}$ . and a leucocytosis, rising to 15,500 per cu. mm. (16 May 1969) six days after admission.

Serial chest films showed an increasing right pleural effusion, and on 13 May a left pleural effusion also appeared. Further Gastrografin examination showed that the oesophageal obstruction was less; but the unusual appearance persisted, with proximal dilatation and a filling defect below the aortic arch. Again no contrast leaked beyond the oesophageal lumen, nor at any time did she show cervical or mediastinal emphysema either clinically or radiologically.

When she was first seen in surgical consultation on 14 May she appeared physically well. In the succeeding days her nocturnal temperature began gradually to settle and the pleural effusions began to clear.

Oesophagoscopy on 20 May showed yellowish purulent fluid in the oesophageal lumen. At 25 cm. from the upper jaw there was a 2-cm. long transverse rent in the mucosa anteriorly. The mucosa of the lower oesophagus and cardia was normal.

*Further progress* The diagnosis was still that of a spontaneous intramural perforation of the oesophageal wall, with fluid causing the submucosal dissection. Although she continued to take a fluid diet,

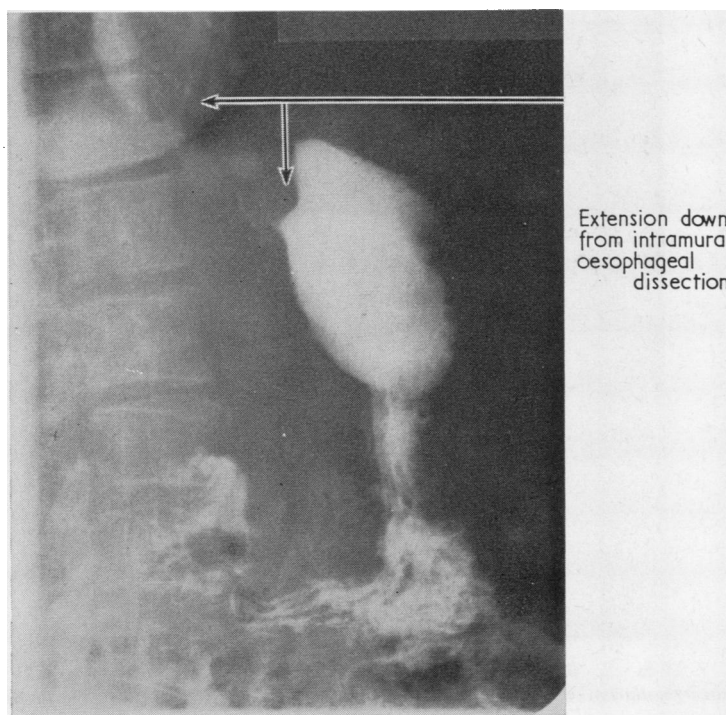


FIG. 4. Case 2. Impression on gastric fundus by extension down from the intramural oesophageal dissection.

her temperature remained raised and her oesophagus was still radiologically abnormal in outline, as shown by repeat barium examinations on 19 and 27 May (Fig. 5). When, therefore, on 30 May there was a return and increase in the bilateral pleural effusions with a temperature of 38° C., thoracotomy was advised and performed that day.

**Operation** The patient was again oesophago-scoped. The mucosal rent at 25 cm. was still seen. The oesophagoscope was passed on to the cardia, which was normal. A wide-bore stomach tube was left in the oesophageal lumen.

A standard *right thoracotomy* was next performed. Some 250 ml. of slightly turbid fluid was aspirated from the right pleural cavity. The mediastinum was turgid and inflamed with ecchymosis along its whole length. There was no surgical emphysema. The mediastinal pleura was incised vertically over a 15-cm. length and the oesophagus was raised by a catheter sling. Tissue fluid freely drained away. The oesophageal wall was more than double normal volume. It was explored at intervals up to the level of the aortic arch, once anteriorly, twice on the right side, opening on to the wide-bore stomach tube, without

convincingly entering an intramural dissection-space.

Each incision in the oesophageal wall was next closed in two layers with interrupted chromic catgut sutures. The mediastinal space was left draining freely into the right pleural cavity. The right chest wall was closed over waterseal drainage by standard technique.

Pleural fluid specimens, taken for culture, proved sterile.

**Left thoracentesis** When a trocar and cannula was next passed into the left pleural cavity and waterseal drainage was established, some 400 ml. of slightly turbid fluid immediately drained out. This, too, proved sterile.

**Post-operative progress** The patient continued with superior vena caval drip therapy for the next week. The left intercostal tube was removed on the second post-operative day and the right intercostal tube on the third post-operative day.

The patient took water by mouth on the fourth post-operative day followed by a fluid diet. She was swallowing normally two weeks after operation and was discharged fit on 26 June 1969. A further check

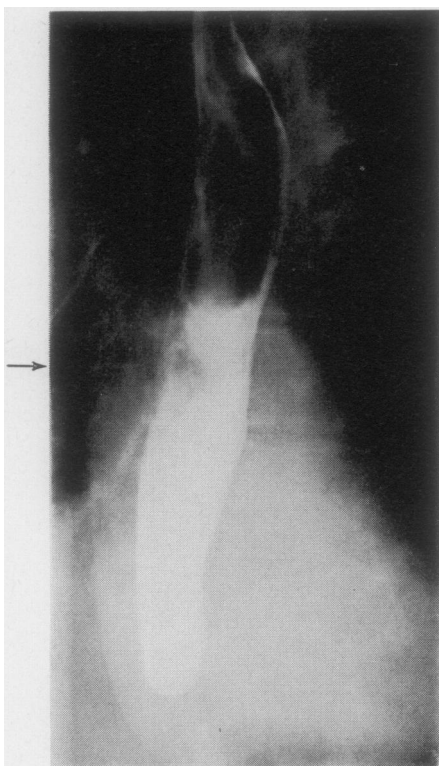


FIG. 5. Case 2. Barium swallow on day 17 shows a mural irregularity on the anterior aspect.

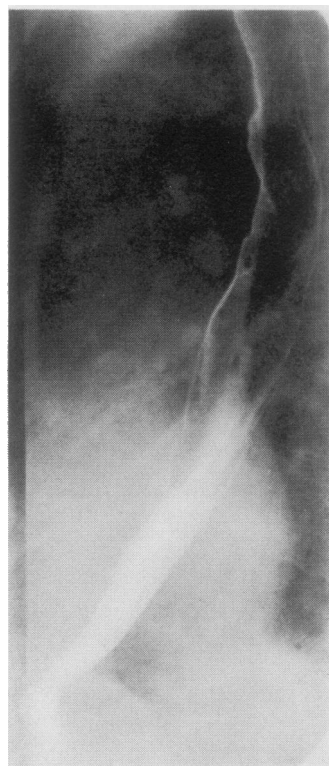


FIG. 6. Case 2. Barium swallow on day 44 shows a return of the oesophageal outline to normal.

barium swallow on day 44 showed a normal oesophageal outline (Fig. 6).

Subsequent oesophageal manometry (courtesy of Mr. I. Lichter, F.R.C.S.) showed the presence of a good inferior oesophageal sphincter. During swallowing the pressure contractions recorded were of moderate amplitude but non-peristaltic.

#### DISCUSSION

It is the clarifying and emphasizing of a broader classification of all aspects of oesophageal perforation, in the light of a more recent clinical experience with spontaneous intramural perforation, that is the burden of this paper. We are unaware of any reports of *acquired* intramural perforation prior to 1957, specifically stated as such, although we do not deny that they may have occurred. Our second case of acquired intramural perforation in 1962 prompted its description as 'dissecting oesophageal abscess' (Lichter and Borrie, 1965).

As a matter of probability it was only a question of time before *spontaneous* intramural perforation of the oesophagus would be seen. The forces which allow of transmural perforation into the mediastinum or pleural spaces could obviously merely breach the oesophageal mucous membrane. The Mallory-Weiss syndrome (Mallory and Weiss, 1929) represents a variant on the 'partial rupture theme', but with emphasis on bleeding and 'haematemesis' rather than on dissection. It seems likely that such a mechanism obtained in Case 1 here reported, where the suppression of an embarrassing sneeze during the swallowing act by an unusually polite gentleman resulted in unusual transmural forces in the oesophagus.

Our interpretation of the events was as follows: The stifling of his sneeze while swallowing food and air created a sudden increased oesophageal pressure that caused a tear of the oesophageal mucous membrane. Later during that afternoon, from the steady, automatic swallowing of saliva (1 ml./min.), and accentuated by his swallowing milk, he had an extensive intramural dissection of the oesophageal mucous membrane aggravated by retching and giving the radiographs pathognomonic appearances of a double-barrelled oesophagus. In this instance, the intramural perforation was 'spontaneous' in type, and as such was the first we had encountered either clinically or in the medical literature at that date (22 September 1967).

The cases of Williams (1957) and of Benjamin and Hanks (1965—Case 3) are noteworthy in

relation to the pathogenesis of this condition generally and of our second case in particular. Williams's patient was an elderly lady and her symptoms of 'fluids sticking halfway down' followed a fall from her bed. Benjamin and Hanks's (1965) patient developed symptoms while on anti-coagulant treatment. Both exhibited radiological signs similar to those shown in our second case and interpreted as 'spontaneous submucosal (intramural) haemorrhage'. Williams, B. (personal communication, 1969) has seen similar appearances in boxers, and his suggestion is that the initial lesion is a submucosal haematoma which may rupture into the lumen, or strip the wall, or resolve spontaneously. Development of communication with the lumen allows of saliva or ingested material gaining access to the mural space and contributing to its extension.

Further stresses of vomiting and retching persisted for some time in both our cases, and such factors could well contribute to the development of the 'full-blown condition'.

*Radiologically*, the diagnosis is made by contrast swallow examination. Water-soluble contrast material, such as Gastrografin (Schering), is preferred, although our cases and those similar ones previously reported have all been demonstrated by the use of barium, without apparent ill effects. The principal sign depends on whether the contrast material enters the dissection-space or not. If it does, it will be seen to lie outside the lumen in a narrow strip, ranging up to a frank duplication ('double-barrelled oesophagus' as in Fig. 1). When contrast *does not* enter the dissection, a filling defect will be seen. When gross, as in Fig. 3, this presents as an extensive deflection of the contrast material with a degree of obstruction above. A longitudinal band-like defect, as shown by Marks and Keet (1968), together with various other bizarre mural defects (see Fig. 5) represent lesser degrees or evidence of resolution. The precise point of entry may or may not be demonstrated at the first examination.

A *delayed film* after one or two hours is most important, for a well-penetrated lateral chest film may reveal persisting contrast in relation to the oesophagus or, as in our Case 1, show drainage from the second lumen. Plain frontal films of the chest are obviously important to demonstrate pleural fluid, mediastinal air or other mediastinal pathology.

It is important to recognize this new variant, Spontaneous Intramural Oesophageal Perforation (SIOP), and incorporate it in the differential diagnosis of sudden chest pain and haematemesis,

particularly with the modern emphasis on 'Coronary Intensive Care' and the understandable likelihood of 'early mis-diagnosis'.

Correct diagnosis depends on (a) careful history and clinical evaluation, (b) an index for suspicion for this lesion and (c) its radiological demonstration by contrast swallow examination.

Exact diagnosis is important because it excludes many other causes, including myocardial infarction, dissecting aneurysm and complete transmural perforation. Further, it allows of reasonable treatment and prognosis.

In our first case, the anatomical site of the perforation was just above the cardia. In our second case, the mucosal breach was seen by oesophagoscopy as a mucosal rent 25 cm. from the upper jaw. The 'solid nature' of her oesophageal shadow and her persisting vomiting of blood made blood the highly likely stripping agent.

Regarding treatment, whilst one cannot prevent the patient swallowing his daily output of saliva, one should ensure that nothing further is given by mouth until the mucous membrane becomes reattached to its muscular layer. In both our patients this was supplemented by superior vena caval supportive therapy. It proved adequate in Case 1 but inadequate in Case 2. In this case the temperature rose after 20 days, because of a sudden increase of bilateral sterile pleural

effusions, and thoracotomy was performed. The release of tissue fluid from a tense but non-purulent mediastinum led to a return of the oesophagus to normal shape and function.

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